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Type II first branchial cyst and sinus excision with preservation of facial nerve and parotid gland

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ABSTRACT

We describe a rare case of type II first branchial cleft sinus and cyst, which was misdiagnosed and mismanaged for many years before being correctly diagnosed and managed. A 3-year-old girl had been seen 16 times over a 3-year period for a recurrent discharging sinus before a correct diagnosis of a first branchial cleft cyst and sinus was made. The patient had an uneventful recovery following total excision of the sinus and cyst with preservation of the facial nerve and parotid gland. We report a novel technique of anatomical and electrophysiological identification of facial nerve and preservation of the facial nerve and parotid gland in type 2 first branchial sinus and cyst deep to the facial nerve trunk involving superficial and deep lobes of the left parotid gland.

Anomalies of the first brachial cleft are rare [1,2,3], with an incidence less than 10% of all branchial arch cleft defects [4] and less than 8% of all branchial anomalies [5] with approximately 200 cases reported in literature [6]. They are typically misdiagnosed and inappropriately managed [7] for several years before being correctly diagnosed and appropriately treated. Data analysis by Triglia et al. shows that the average delay between initial presentation and adequate treatment was 3.5 years The delay in diagnosis can result in unnecessary investigations and iatrogenic injuries and unnecessary anguish for the patient [8,9]. Traditionally type 2 first branchial cyst and sinus excision will involve either superficial or total parotidectomy with some injury to the facial nerve, we describe the novel technique of total excision of the cyst and sinus with total preservation of both the facial nerve and the parotid gland.

1. Case report

A 6-month-old girl presented in January 2009 with a left submandibular abscess which was drained uneventfully. However, the lesion itself persisted and continued to discharge small amounts of watery pus on a regular basis (Fig. 1A). The patient had been seen by her general practitioner 4 times prior for a non-resolving abscess and cellulitis with courses of flucloxacillin and doxycyclin. The patient then had a further 12 recurrent hospital consultations for a possible atypical tuberculous lymph node infection in the left submandibular region, where the presenting complaint was of an intermittent watery pus discharge accompanied with a painful lump in the region of the left parotid gland.

Each hospital visit was compounded by the abscess having healed, with no obvious discharge, thus no active investigation were pursued. It was after the patients' twelfth hospital appointment that imaging was considered for a possible diagnosis of a first branchial cleft cyst.

Initial imaging with an ultrasound scan revealed that from the visible puckered scar there was a 4 mm diameter tract running deep passing lateral to the left submandibular gland. It can be traced for about 1.5 cm and ending in the deep part of left parotid gland in a 2 cm long and about 8 mm heterogenous well defined cyst with vascularity on Color Doppler scan (Fig. 2A and B).

A magnetic resonance imaging (MRI) scan under general anesthesia revealed a left neck sinus tract, adjacent to the left submandibular gland and running deep to the superficial level of the left parotid gland ending into a small 9.9 mm × 9.5 mm focal

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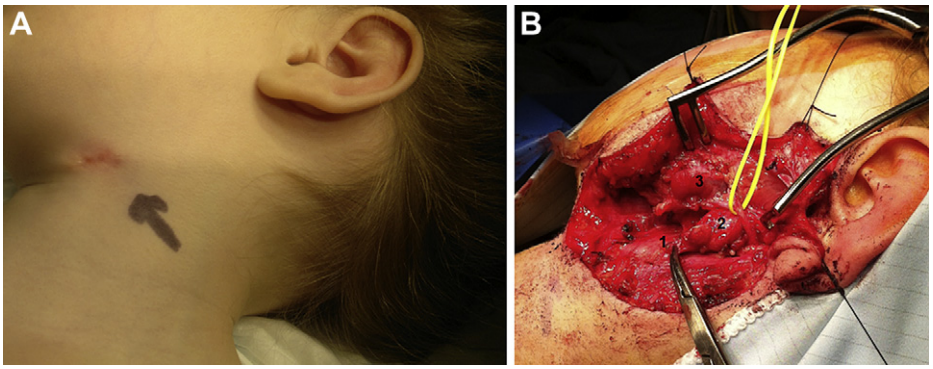

Fig. 1. A and B, Clinical and operative photographs. Cyst [1], Sinus Tract [2] and isolation and preservation of the facial nerve (sling) and the parotid gland [3].

area of high T2 signal region which was suggestive of a first branchial cleft cyst and sinus (Fig. 2C and D).

Following discussion with the Ear Nose and Throat (ENT) surgeons an operation was scheduled. At operation total excision of first branchial sinus and cyst was carried out with preservation of the facial nerve using, both visual anatomical and electrophysiological identification, and both parts of the left parotid gland (Fig. 1B).

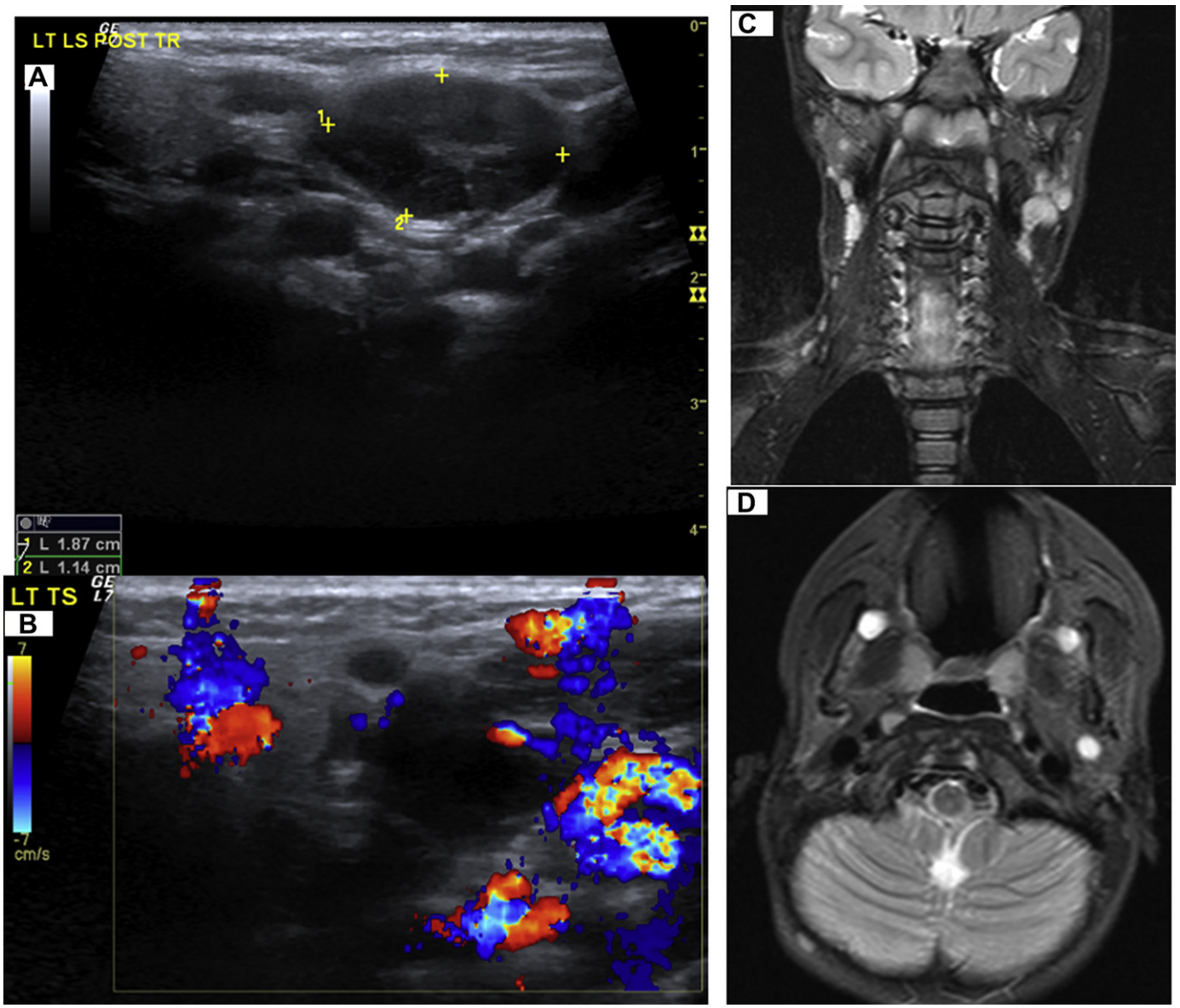

Fig. 2. A, Ultrasound 2B Color Doppler and; C&D, MRI scans showing the location and extent of the sinus and the cyst.

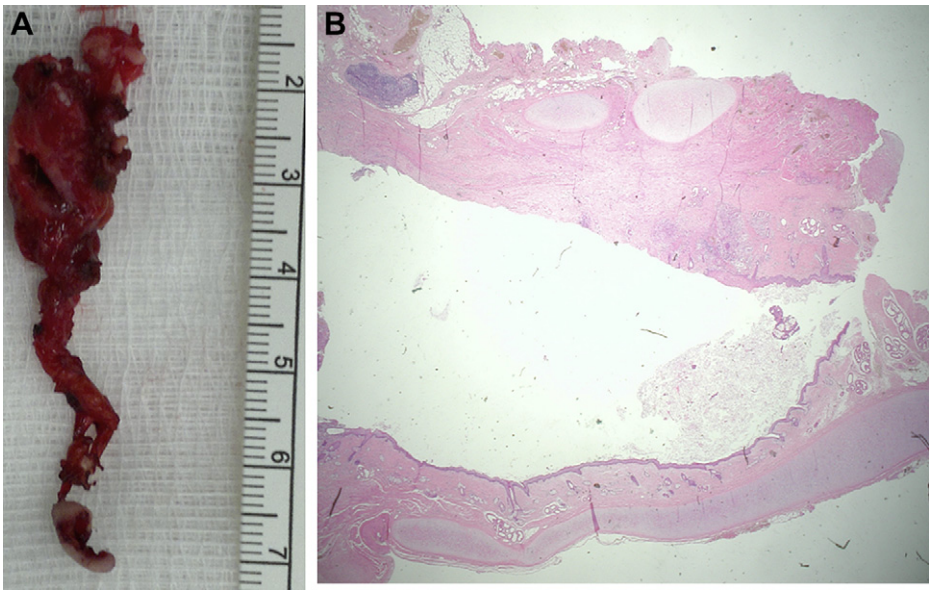

Fig. 3. A, Excised sinus and cyst. B, Histology showing squamous epithelium, adnexal structures, cartilage and inflammation.

The cyst and sinus were accessed using a modified Blair's incision, where an incision was made in the preauricular crease and extending it along the margins of the ramus and body of the mandible.

The cyst and sinus was successfully excised without causing facial nerve damage. There was some mild residual post-operative weakness in the facial nerve distribution due to intra-operative stretching and the patient was discharged the following day with no complications.

Histological examination of the excised the cyst and sinus confirmed intact lining of squamous epithelium with dermal-like tissue containing adnexal structures and a cartilage in the deeper plane consistent with first branchial remnants (Fig. 3A and B).

At outpatient appointment one month later, where there was no evidence of a reoccurrence and a good post-operative outcome, the patient was discharged with no further follow up.

2. Discussion

During the fifth week of development, branchial arches appear on the lateral side of the embryonic face, which then protrude into the early foregut [10], thus helping to establish the embryonic neck [11]. The ridges that are formed by this action are separated by four pairs of ectodemal depressions; also know as the branchial clefts and the internally formed pouches of endodermal origin. The arches formed by this process give rise to the formation of the various head and neck structures. The first arch, also known as the mandibular arch, typically appears around day 22 and forms the skin of the lower part of the face, malleus, incus, mandible, maxilla, and the muscles of mastication [10].

The classification of first brachial cleft anomalies was produced by Work in 1972 and is based on the histological features present. Type I typically present as a cystic mass and are ectodermal in nature, where as type II typically present as either a sinus, cyst or fistula or a combination of these lesions, and are of ectodermal and mesodermal origins [8]. Histological samples from our patient confirmed that it was indeed a branchial cleft cyst and sinus. The histopathology report showed that the sinus in this case was lined with dermal like tissue with sweat glands, adnexal structures and cartilage, in keeping with a type 2 first branchial cleft anomaly.

First branchial cleft anomalies arise due to the incomplete closure of the ectodermal portion of the first branchial cleft. It is the degree of incomplete closure, which determines whether the defect would be a sinus, cyst, fistula or a combination [8]. In the study of 39 cases by Triglia et al., all of the first branchial cyst anomalies were located in a triangular region of the neck. This triangle is formed by the external auditory canal, medial to the intertragic notch (which is a remnant of the first groove of the first cleft), to the base of the triangle, formed by a line between the chin and the mid point of the hyoid bone, with superior border following the lines of the mandible [8]. In our patient, the abnormality was in the submandibular region. Anomalies of the first branchial cleft are typically located close to the superficial lobe of the parotid gland.

Appropriate imaging is required preoperatively in the surgical planning of the excision. Ultrasound was helpful to an extent in identifying the sinus tract; however the quality of the scan and extraction of detail is very user dependent. In our patient, subsequent MRI scan was carried out under general anesthesia for more detailed information. In a study by Mukherji, ST et al., the radiological team looked at the use of computed tomography (CT) scan and MRI scan in the evaluation of first branchial anomalies. They showed that in the 11 patients seen over a 10 year period, CT scans was preferred over MRI scans due to the visualization of bony detail and also the better capability to determine the cystic nature of the lesion [11].

As reported by Issacson and Martin in 2000, the use of electrophysiological means for the identification of the facial nerve resulted in successful excisions of first branchial cleft cysts with no residual facial weakness over a 9-year period, and with all having a better cosmetic outcome [12]. Magdy et al. outline 18 cases involving first branchial cleft anomalies where electrophysiological facial nerve monitoring was used in all 18 cases [13].

3. Conclusion

It is important to keep in mind the possibility of a branchial cleft anomaly in recurring infections of the ear and neck, especially if the prescribed treatments do not resolve the problem. Early imaging is warranted in these types of patients especially when there is a high degree of suspicion. The choice of imaging is dependent on the age

of the patient. Early intervention reduces the anguish for the patient and reduces the risk of reoccurrence and improper treatment. The only appropriate management of a first branchial cleft anomaly is careful surgical excision. Type 2 first branchial anomalies lie very close to the facial nerve and are therefore at higher risk of iatrogenic injury, resulting in associated facial palsies. Per operative identification of the facial nerve by anatomical and/or by electrophysiological means is absolute. Meticulous dissection around the sinus leading to the cyst and remaining closer to the cyst allows preservation of the superficial and deep lobes of the parotid gland as well.

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